

# Not all Sicca is Sjogrens and Not all Sjogrens is Sicca



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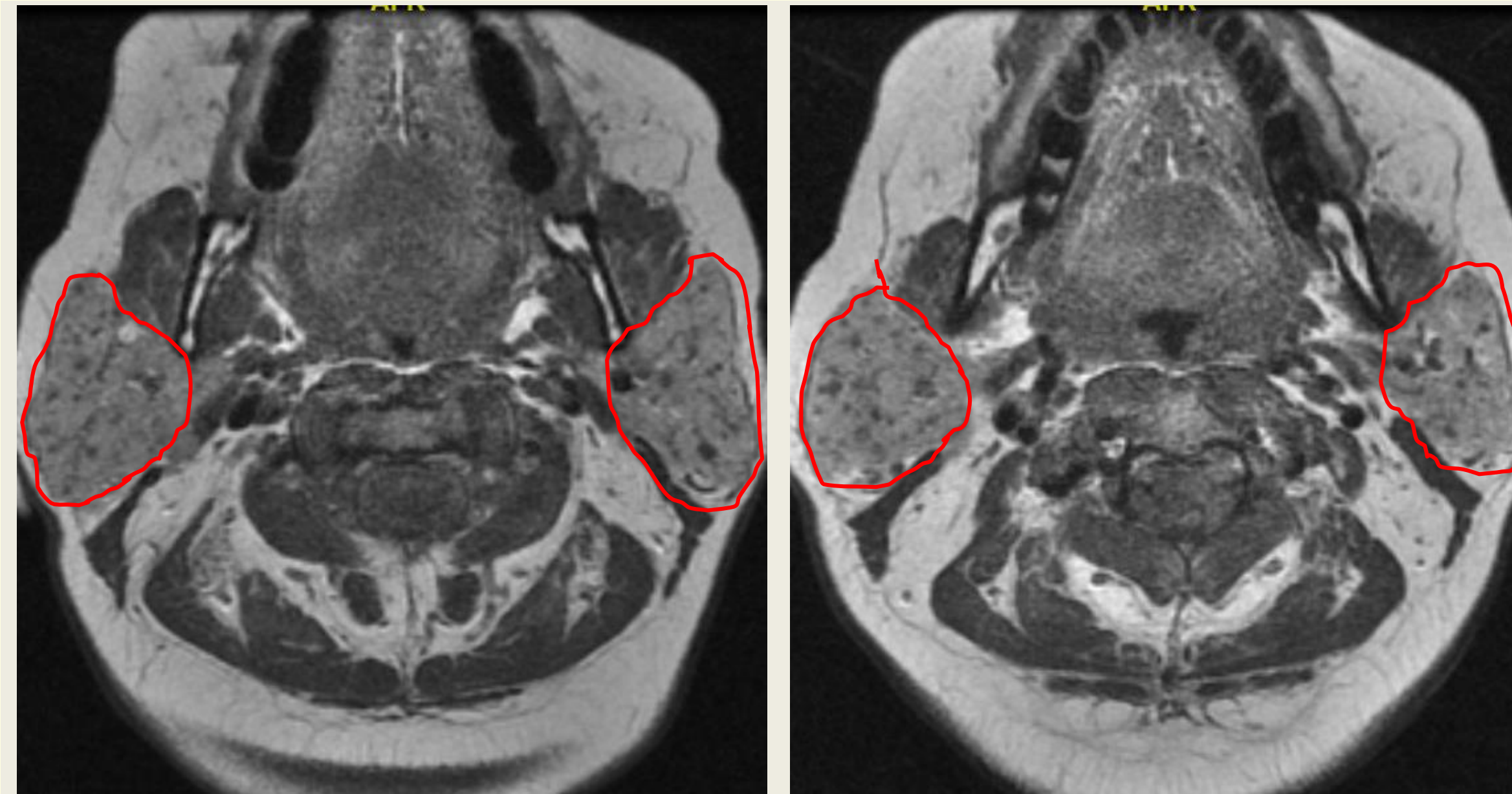
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## Introduction

- Symptoms of dry eyes or dry mouth otherwise known as sicca symptoms are not always present in patients with Sjogrens Syndrome
- Approximately 20% of patients with Sjogrens Syndrome don't have sicca symptoms
- Diagnosis of Sjogrens Syndrome is a clinical diagnosis suspected in patient with sicca symptoms or with clinical suspicion of end organ involvement. Diagnosis is supported by proof of autoimmunity, salivary gland involvement, salivary gland biopsy or objective evidence of dry mouth by sialometry or dry eyes by schirmers test or vital dye stain scoring.

## Case Report

- 62-year-old female presents to the rheumatology clinic with complaints of left sided facial weakness.
- Facial weakness was of sudden onset and has persisted for the past 2 months.
- Received empiric treatment for Bell's palsy with low to medium dose prednisone for 3 weeks and 1 week acyclovir without improvement.
- History of one previous episode of bells palsy that self resolved in 1995.
- PMHx: bilateral SNHL, HTN. Home Rx: Olmesartan, Aspirin
- Lifetime nonsmoker, native from Philippines.
- Physical exam: vitals WNL. Bilateral facial fullness and complete left sided facial hemiparesis including upper and lower face with inability to wrinkle her forehead. Extraocular muscles were intact. No oral or nasal lesions. No palpable cervical or axillary adenopathy. Chest and cardiovascular exam was unremarkable. There was no palpable organomegaly on abdominal exam. Strength of upper and lower extremities was 5/5 bilaterally.
- Workup remarkable for abnormal MRI shown here and elevated ANA with positive SSA/Ro Antibody.
- Based on clinical suspicion and supported by imaging and serology, patient is diagnosed with Facial nerve palsy secondary to Sjogrens Syndrome. After thorough workup and evaluation, patient started on immunosuppressive therapy with mycophenolate mofetil and prolonged steroid taper.
- Diagnosis can be confirmed with salivary gland biopsy.



## MRI Brain with and without contrast

T1-Weighted images in sequential order craniocaudally (right to left image) shows enlarged parotid glands bilaterally (outlined in red), more pronounced on right side with nodular ductal dilatations and mixed calcifications. This MRI also showed contrast enhancement of the tympanic segments of bilateral facial nerves, compatible with facial neuritis.

## Remarkable Labs

Remarkable Labs	
Serologies	ANA 1:80 speckled SSA/Ro positive >8 A.I.
HIV	Negative
Lyme Serology	Negative
Hepatitis Panel	Negative for HBV core ab or HBSA, HCV
Inflammatory markers	ESR, CRP WNL
SPEP	Negative for gammopathy

## Discussion

- This patient presented with recurrent and now persistent left facial nerve palsy
- When evaluating a patient with facial nerve palsy it is important to consider different causes like Stroke, Otitis Media, Sarcoidosis, HSV, HIV, Lyme disease, Guillain Barre syndrome or a local tumor. Ultimately most patients are determined to have idiopathic facial nerve palsy (Bell's palsy).
- The most common rheumatic disease associated with facial nerve palsy is Sjogrens Syndrome.
- Despite its classic association with dry eyes and dry mouth syndrome, approximately 20% of patients with Sjogrens Syndrome (SS) do not have sicca symptoms. Moreover, only 10% of patients with clinically significant dry eye syndrome have SS.
- SS can present with a myriad of other systemic manifestations including cranial nerve neuropathies as our patient. Interestingly as many as 6 Cranial nerves can be involved simultaneously. Like our patient with h/o SNHL (CNVIII) and now facial nerve palsy (CN VII).
- Currently Classification criteria for SS does not include consideration of abnormal imaging but experts stipulate it can be suspected if certain particular findings on MR, Ultrasound, CT or salivary scintigraphy or sialography.
- Consider a clinical diagnosis of SS if 3 out these 4 findings:
  - ANA 1:320, RF+, SSA+
  - Positive Lip gland biopsy showing focal lymphocytic sialadenitis with >1 focus score
  - Evidence of dry eyes (schirmers or abnormal ocular surface staining)
  - Evidence of salivary gland involvement (abnormal imaging or by sialometry)

## References:

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